

acid sequence of the recombinant α -glucosidase produced in the milk of mice was shown to be the same as that of α -glucosidase precursor from human urine as published by Hoefsloot et al., *EMBO J.* 7:1697-1704 (1988) which starts with AHPGRP.

While the foregoing invention has been described in some detail for purposes of clarity and understanding, it will be clear to one skilled in the art from a reading of this disclosure that various changes in form and detail can be made without departing from the true scope of the invention. All publications and patent documents cited in this application are incorporated by reference in their entirety for all purposes to the same extent as if each individual publication or patent document were so individually denoted.

What is claimed is:

1. A transgenic nonhuman mammal having a transgene comprising:

a promoter and enhancer from the same mammary gland specific gene;

a secretory DNA segment encoding a signal peptide functional in mammary secretory cells of the transgenic nonhuman mammal, and

a recombinant DNA segment encoding acid α -glucosidase operably linked to the secretory DNA segment to form a secretory-recombinant DNA segment, the secretory-recombinant DNA segment being operably linked to the promoter and enhancer, and wherein the secretory DNA segment is an acid α -glucosidase secretory DNA segment or is from the same mammary-gland specific gene as the promoter and enhancer;

wherein the transgene, in an adult form of the nonhuman mammal or a female descendant of the nonhuman mammal, expresses the secretory-recombinant DNA segment in the mammary secretory cells to produce acid α -glucosidase that is processed and secreted by the mammary secretory cells into milk in a recoverable amount with α -glucosidase catalytic activity.

2. The transgenic nonhuman mammal of claim 1, wherein the concentration of the acid α -glucosidase in the milk is at least 100 $\mu\text{g/ml}$.

3. The nonhuman transgenic mammal of claim 1, wherein the secretory DNA segment is an acid α -glucosidase secretory DNA segment.

4. The transgenic nonhuman mammal of claim 1, wherein the human acid α -glucosidase is secreted into milk in a form that can be taken up by muscle cells.

5. The nonhuman transgenic mammal of claim 1, wherein the acid α -glucosidase is human.

6. The nonhuman transgenic mammal of claim 5, that is a mouse or rabbit.

7. The nonhuman transgenic mammal of claim 6, wherein the recombinant DNA segment is cDNA.

5 8. The nonhuman transgenic mammal of claim 6, wherein the recombinant DNA segment is genomic.

9. The nonhuman transgenic mammal of claim 6, wherein the recombinant DNA segment is a cDNA-genomic-DNA hybrid.

10 10. A method for producing acid α -glucosidase, the method comprising:

recovering milk from the adult form of the transgenic nonhuman mammal of claim 1 or its female descendant, wherein said milk contains a recoverable amount of acid α -glucosidase with catalytic activity.

15 11. The method of claim 10, further comprising incorporating the milk into a food product.

12. The method of claim 10, further comprising purifying the acid α -glucosidase from the milk.

20 13. The method of claim 12, wherein the acid α -glucosidase is purified to at least 95% pure from other components of the milk.

25 14. The method of claim 13, further comprising mixing the acid α -glucosidase with a pharmaceutical carrier for intravenous, intradermal, intramuscular or oral administration.

30 15. Milk from the transgenic nonhuman mammal of claim 1, the milk comprising human acid α -glucosidase in a recoverable amount.

16. The milk of claim 15, wherein the concentration of the human acid α -glucosidase is at least 100 μ g/ml.

35 17. A composition comprising human acid α -glucosidase with catalytic activity and capacity to be taken up by muscle cells in a patient and milk of the nonhuman transgenic mammal of claim 1.

40 18. A pharmaceutical composition for parenteral administration to a human patient comprising human acid α -glucosidase with catalytic activity and in a therapeutically effective dosage to treat a patient suffering from Pompe's disease; and a pharmaceutical carrier, the composition being free of other human proteins present in its natural environment.

45 19. The pharmaceutical composition of claim 18, wherein the pharmaceutical carrier is for intravenous administration.

20. The pharmaceutical composition of claim 18, wherein the human acid α -glucosidase is purified to homogeneity.

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